CHAPTER 7

Episclera and Sclera

Applied anatomy

The scleral stroma is composed of collagen bundles of varying size and shape that are not as uniformly orientated as in the cornea. The inner layer of the sclera (lamina fusca) blends with the suprachoroidal and supraciliary lamellae of the uveal tract. Anteriorly the episclera consists of a dense, vascular connective tissue which lies between the superficial scleral stroma and Tenon capsule. The three vascular layers that cover the anterior sclera are as follows:

1. **The conjunctival vessels** are the most superficial; arteries are tortuous and veins straight.
2. **The vessels within Tenon capsule** are straight with a radial configuration (Fig. 7.1a). In episcleritis, maximal congestion occurs within this vascular plexus (Fig. 7.1b), which can be manually moved over the sclera. Tenon capsule and the episclera are infiltrated with inflammatory cells, but the sclera itself is not swollen. Instillation of topical phenylephrine will cause blanching of the conjunctiva, and to a certain extent the Tenon vessels, allowing visualization of the underlying sclera.
3. **The deep vascular plexus** lies in the superficial part of the sclera and shows maximal congestion in scleritis (Fig. 7.1c). There is also inevitably some engorgement of the superficial vessels, but this should be ignored. Topical phenylephrine has no effect on the engorgement of these vessels. Examination in daylight is important to localize the level of maximal injection. The scleral stroma is largely avascular.

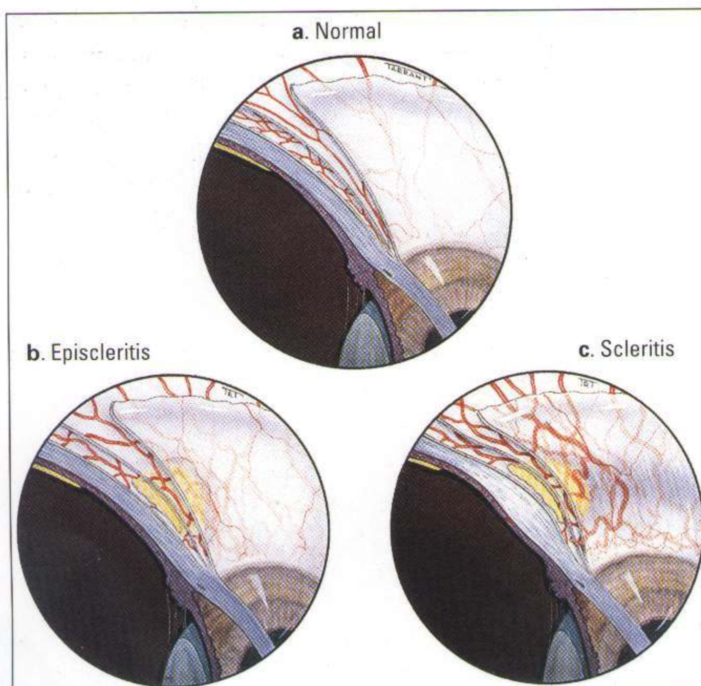


Fig. 7.1
Anatomy of the anterior vascular coats in relation to episcleritis and scleritis

Episcleritis

Episcleritis is a common, benign, self-limiting and frequently recurrent disorder which typically affects young adults. It may on occasion be associated with underlying systemic disease but never progresses to true scleritis. Episcleritis may be (a) *simple* or (b) *nodular*.

Clinical features

1. **Presentation** is with unilateral redness associated with mild discomfort, tenderness and watering.
2. **Signs**
   *a. **Simple episcleritis***, the commonest type, is characterized by sectoral (Fig. 7.2), or less commonly, diffuse

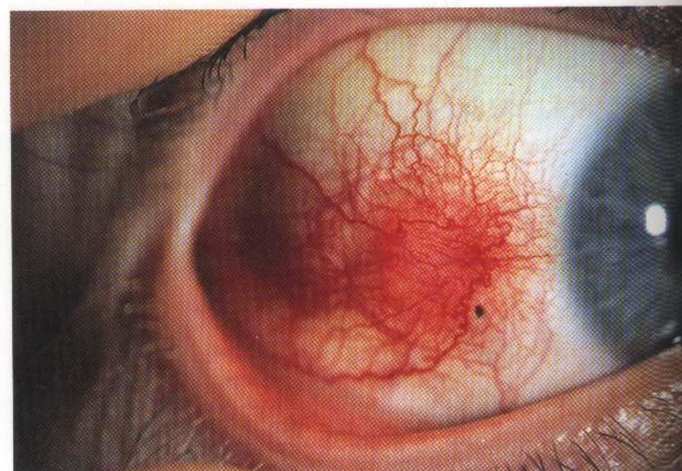


Fig. 7.2
Simple sectoral episcleritis

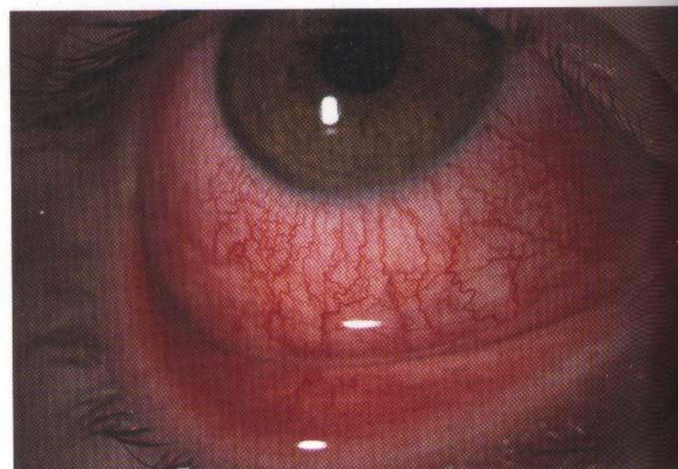


Fig. 7.3
Simple diffuse episcleritis

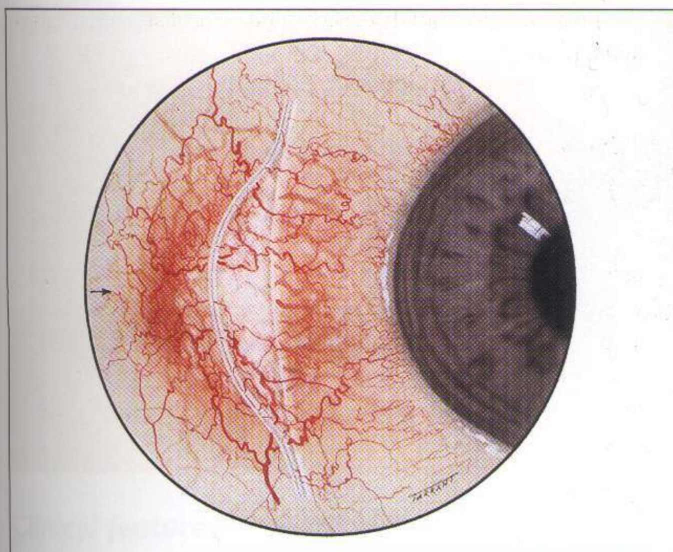


Fig. 7.4
Nodular episcleritis (Courtesy of P. Watson)

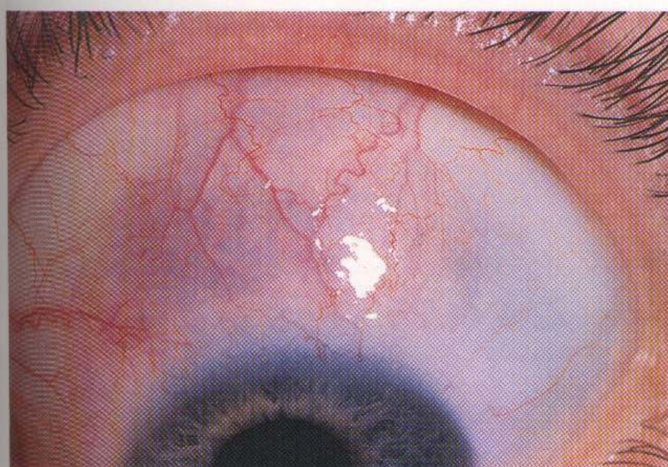


Fig. 7.5
Scleral translucency following recurrent episcleritis

redness (Fig. 7.3). It usually resolves spontaneously within 1–2 weeks.

b. Nodular episcleritis is characterized by a localized, raised, congested nodule which takes longer to resolve.
- A thin slit-lamp section shows that the anterior scleral surface is not raised, indicating that the sclera is not swollen (Fig. 7.4).
- Following recurrent attacks, the superficial scleral lamellae may become rearranged into more parallel rows, making the sclera appear more translucent (Fig. 7.5). This should not be mistaken for scleral thinning.

Treatment

This is not always required unless symptoms dictate otherwise.

1. **Simple lubricants** or vasoconstrictors suffice in most mild cases.
2. **Topical steroids** may be helpful but their use may result in recurrences. Frequent intensive instillation on a short-term pulsed basis is recommended.
3. **Oral non-steroidal anti-inflammatory drugs** (NSAIDs) such as flurbiprofen 100 mg t.i.d. for a few days may be required for severe recurrent or prolonged inflammation.

Scleritis

Scleritis is characterized by oedema and cellular infiltration of the entire thickness of the sclera. It is much less common than episcleritis and covers a spectrum ranging from trivial self-limiting episodes to a necrotizing process that may involve adjacent tissues and threaten vision.

Causes and associations

1. **Systemic associations** are present in about 50% of patients. Rheumatoid arthritis is by far the most common, followed by Wegener granulomatosis, relapsing polychondritis and polyarteritis nodosa (*see* Chapter 20).
2. **Surgically induced** scleritis follows ocular surgery. The exact aetiology is unknown but it has a strong association with underlying systemic disease and is more common in females. It typically presents within 6 months postoperatively as a focal area of intense inflammation and necrosis adjacent to the surgical site (Figs 7.6, 7.7).
3. **Infectious** scleritis is most frequently caused by spread of infection from a corneal ulcer. It may also be associated with trauma or follow excision of a pterygium with adjunctive beta irradiation or mitomycin C. The most frequent causative organisms are *Pseudomonas aeruginosa*, *Strep. pneumoniae*, *Staph. aureus* and Varicella zoster virus. Pseudomonal infection is difficult to treat and carries a poor prognosis. Fungal scleritis is very rare (Fig. 7.8).

Anatomical classification

This is based on the primary anatomical site of inflammation and associated vascular changes.

1. **Anterior scleritis** (98%)
   a. Non-necrotizing (85%): diffuse or nodular.
   b. Necrotizing (13%): with or without inflammation.
2. **Posterior scleritis** (2%)

Anterior non-necrotizing scleritis

Clinical features

1. **Presentation** is similar to episcleritis although discomfort may be more severe.


Fig. 7.6
Scleritis following scleral buckling for retinal detachment

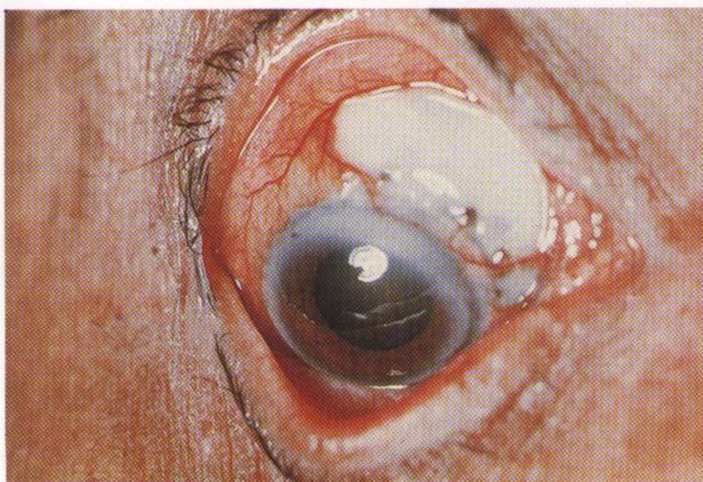
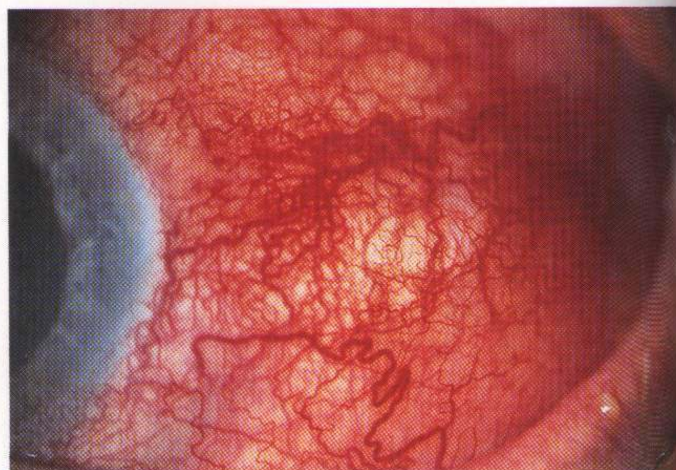
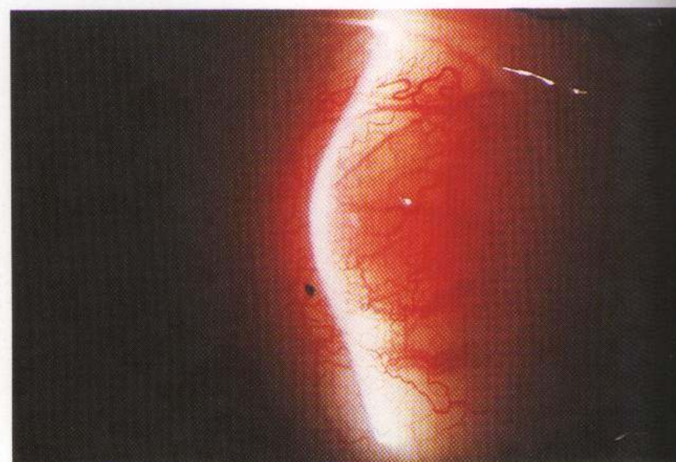

Fig. 7.7
Scleritis following glaucoma filtration surgery


Fig. 7.8
Fungal scleritis (Courtesy of C. Barry)

2. Signs

a. ***Diffuse scleritis*** is characterized by widespread inflammation involving a sector or the entire anterior sclera. Distortion of the normal radial vascular pattern is characteristic (Fig. 7.9). The condition is relatively benign and neither progresses to the nodular type nor becomes necrotizing.


Fig. 7.9
Diffuse non-necrotizing anterior scleritis


Fig. 7.10
Nodular non-necrotizing anterior scleritis

b. ***Nodular scleritis*** (Fig. 7.10) may, on cursory examination, resemble nodular episcleritis. However, the scleral nodule cannot be moved over the underlying tissue. Nodular disease is of intermediate severity with an overall incidence of visual impairment of about 25%.

Treatment

1. **Oral NSAIDs** such as flurbiprofen 100 mg t.i.d. or meloxicam 7.5 mg t.i.d. is the initial treatment.
2. **Oral prednisolone** 40–80 mg daily may be required as short-term therapy in patients resistant to or intolerant of NSAIDs.
3. **Combined therapy** with an NSAID and lower dose of steroid may be effective in the few patients who respond inadequately to either drug alone.

4. **Subconjunctival steroid injection** with triamcinolone acetonide (40 mg/ml) is a safe and effective alternative to systemic therapy but only for *non-necrotizing disease*.

Anterior necrotizing scleritis with inflammation

This is the most severe and distressing form of scleritis. It is bilateral in 60% of cases but involvement is not necessarily simultaneous. Most patients have an associated systemic vascular disease with a mortality rate of 25% within 5 years of the onset of scleritis. The visual prognosis is also poor.

Clinical features

1. **Presentation** is with gradual onset of pain and localized redness. The pain becomes severe and persistent and radiates to the temple, brow or jaw. It frequently interferes with sleep and responds poorly to analgesia.
2. **Signs** (in chronological order)
   - Congestion of the deep vascular plexus (Fig. 7.11).
   - Vascular distortion and occlusion, with resultant avascular patches (Fig. 7.12).
   - Scleral necrosis which may be associated with overlying conjunctival ulceration (Fig. 7.13).
   - Gradual spread of necrosis which may coalesce with other separate necrotic areas (Fig. 7.14).
   - Upon resolution a bluish tinge appears due to increased visibility of underlying uvea secondary to scleral thinning (Fig. 7.15).

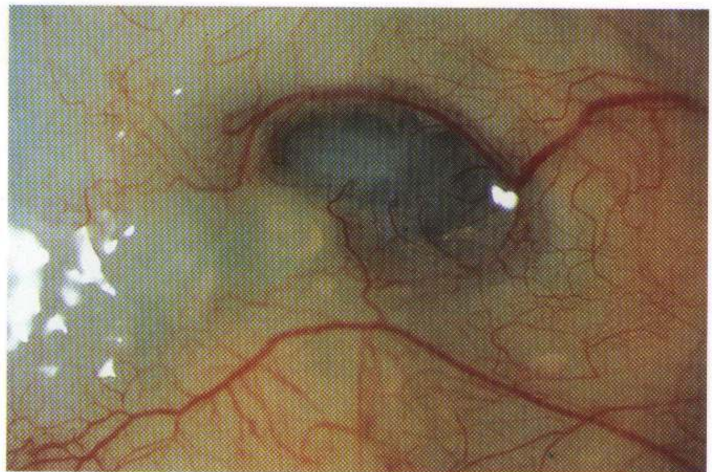


Fig. 7.13
Patch of scleral necrosis in anterior necrotizing scleritis with inflammation

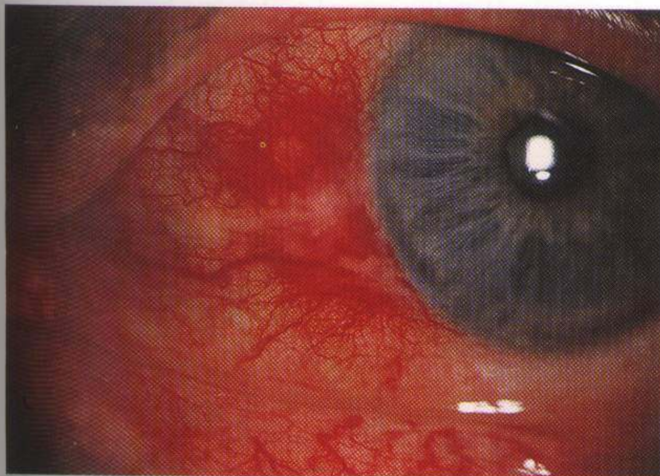


Fig. 7.11
Deep vascular congestion in early anterior necrotizing scleritis with inflammation

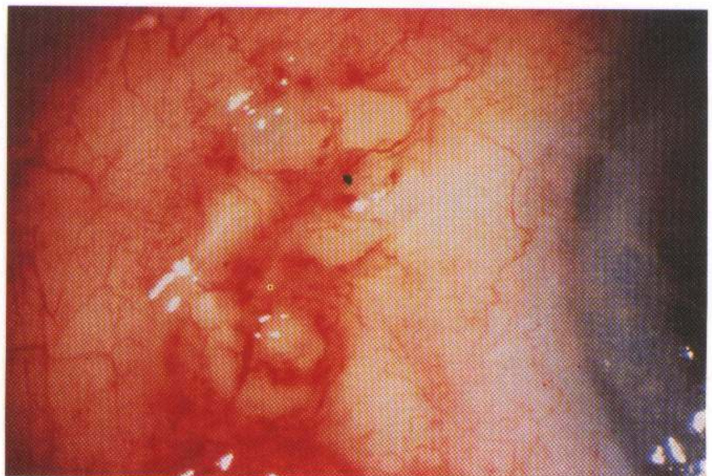


Fig. 7.14
Advanced anterior necrotizing scleritis with inflammation (Courtesy of P. Watson)

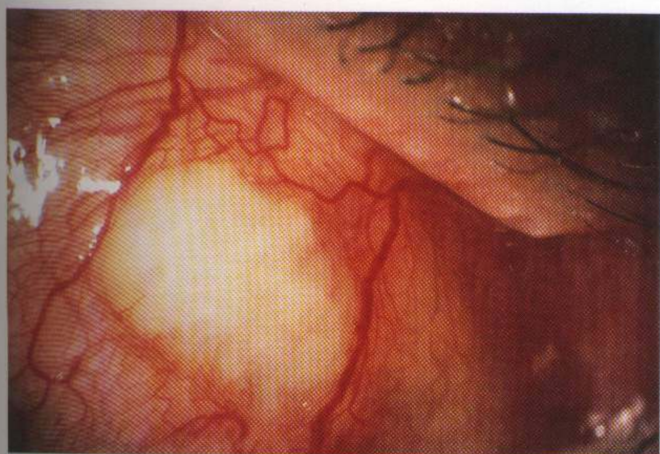
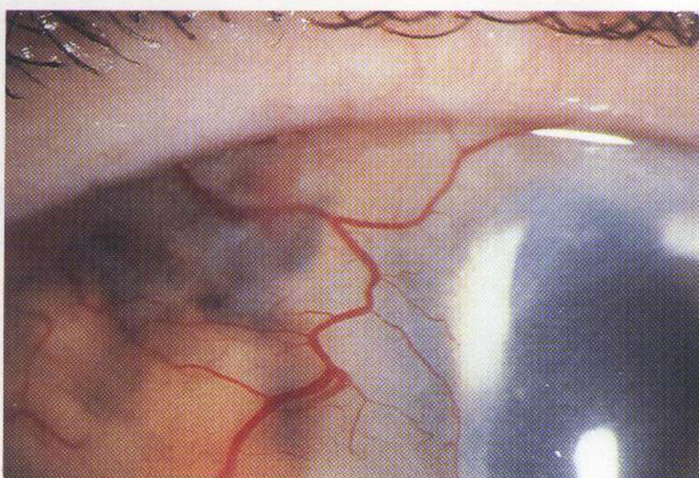


Fig. 7.12
Avascular patch in anterior necrotizing scleritis with inflammation (Courtesy of P. Watson)

Complications

1. **Staphyloma formation** (Fig. 7.16), and occasionally perforation, may occur secondary to severe scleral

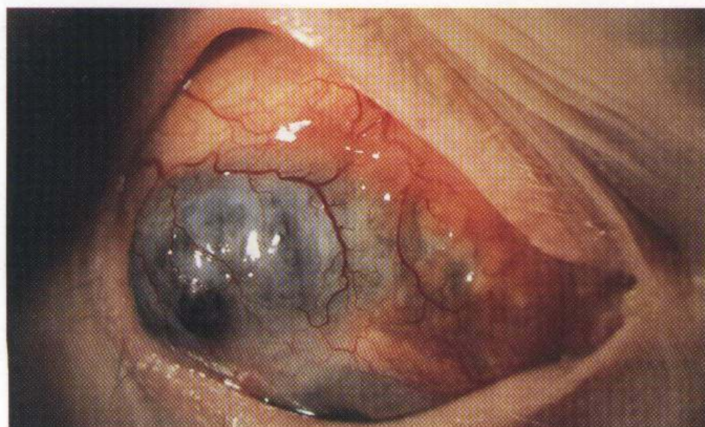

Fig. 7.15
Uveal visibility due to scleral thinning in resolved anterior necrotizing scleritis with inflammation

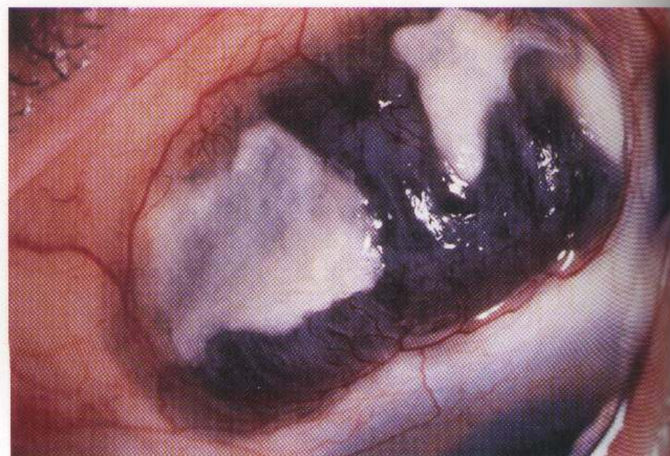
thinning, particularly if the intraocular pressure is elevated.

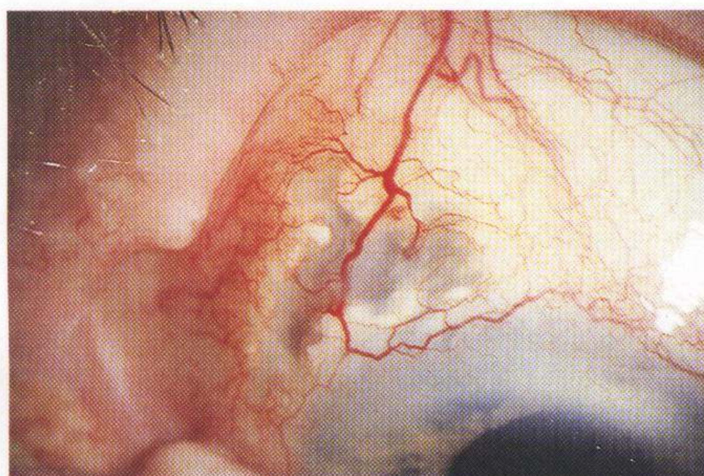
2. **Anterior uveitis** reflects extension of inflammation to the uvea in severe disease. Long-standing uveitis may result in secondary cataract, glaucoma and macular oedema. It is therefore important to detect and treat uveitis promptly. In general, the prognosis is poor with a high incidence of visual impairment.

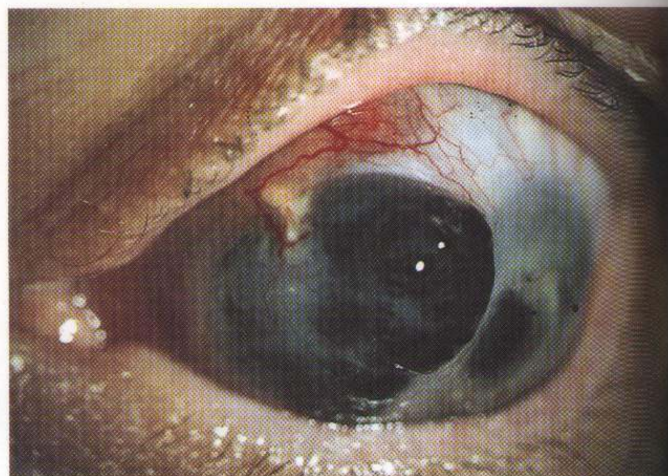
Treatment

1. **Oral prednisolone** 60–120 mg daily for 2–3 days usually has a dramatic effect on the severity of pain, which is an important indicator of active disease. The dose can then be tapered accordingly.
2. **Immunosuppressive agents** (cyclophosphamide, azathioprine or cyclosporin) may be necessary in steroid-resistant cases.


Fig. 7.16
Scleral staphyloma due to anterior necrotizing scleritis with inflammation


Fig. 7.18
Advanced scleromalacia perforans


Fig. 7.17
Early scleromalacia perforans


Fig. 7.19
Scleral staphyloma in severe scleromalacia perforans

3. **Combined therapy** (pulsed intravenous methylprednisolone 500–1000 mg and cyclophosphamide 500 mg) is reserved for the minority of patients who fail to resolve with oral therapy or those who present with established scleral necrosis.

Anterior necrotizing scleritis without inflammation

Also known as scleromalacia perforans, this typically occurs in women with long-standing rheumatoid arthritis and is usually bilateral.

1. **Signs** (in chronological order)
   - Asymptomatic yellow necrotic scleral patches in uninflamed sclera.
   - Enlargement, spread and coalescence.
   - Progressive exposure of underlying uvea as a result of scleral thinning (Figs 7.17, 7.18).
   - Staphyloma formation (Fig. 7.19) may occur but spontaneous perforation is rare unless the intraocular pressure is elevated.
2. **Treatment** is ineffective.

Posterior scleritis

Posterior scleritis is uncommon and often misdiagnosed because it may present with a range of clinical findings which may be confused with other inflammatory and neoplastic conditions. The condition affects women twice as often as men and one-third of patients are under 40 years of age at presentation. Patients over 50 years are at increased risk of harbouring a systemic disease and suffering visual loss. Two-thirds of cases have unilateral involvement. There is no correlation between unilateral and bilateral cases and systemic disease or visual loss. In general the visual prognosis is guarded and about one-third of patients develop visual impairment to some degree.

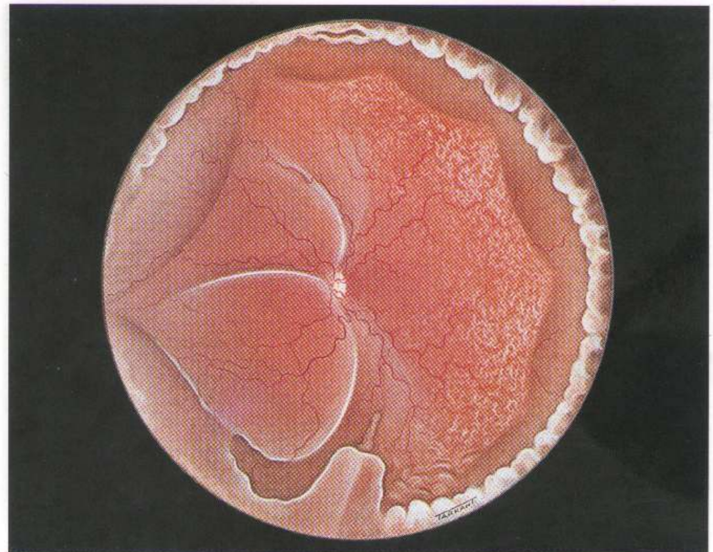


Fig. 7.21
Exudative retinal detachment and ring choroidal detachments in posterior scleritis

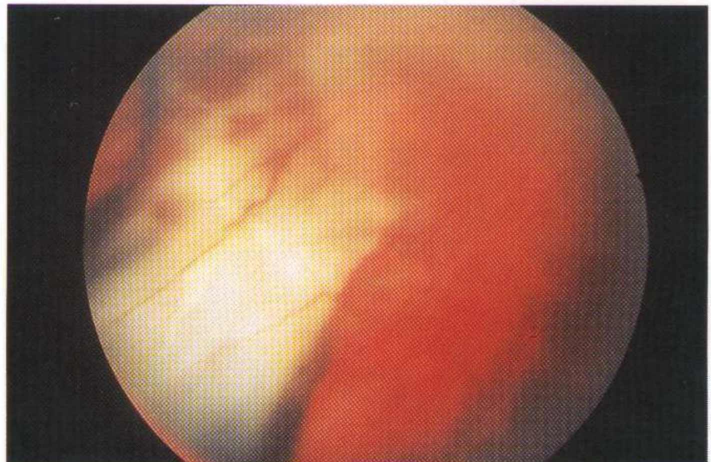


Fig. 7.22
Subretinal lipid exudation in posterior scleritis (Courtesy of P. Watson)

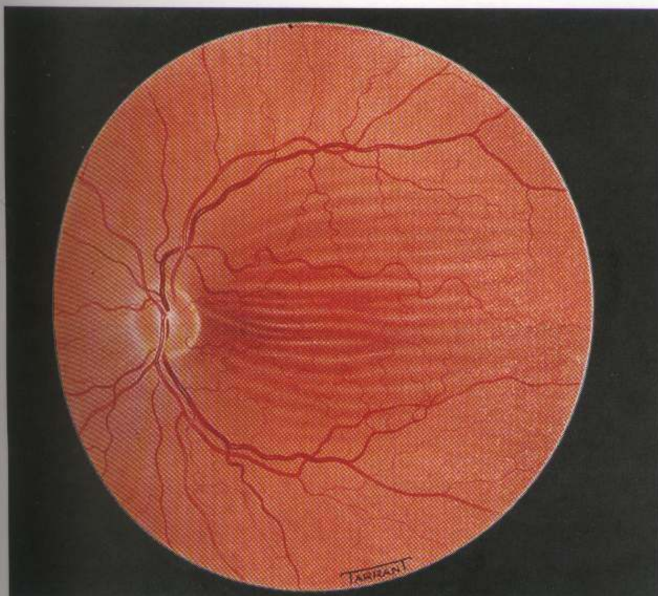


Fig. 7.20
Choroidal folds in posterior scleritis

1. **Presentation** is variable and depends on the exact site of involvement. The most common symptoms are pain and visual impairment.
2. **Signs**
   a. ***External*** signs include lid oedema and fullness, proptosis and ophthalmoplegia. Associated anterior scleritis is present in about one-third of cases.
   b. ***Fundus*** findings include disc swelling, macular oedema, choroidal folds (Fig. 7.20), exudative retinal



Fig. 7.23
Ultrasonogram in severe posterior scleritis showing scleral thickening and fluid in sub-Tenon space

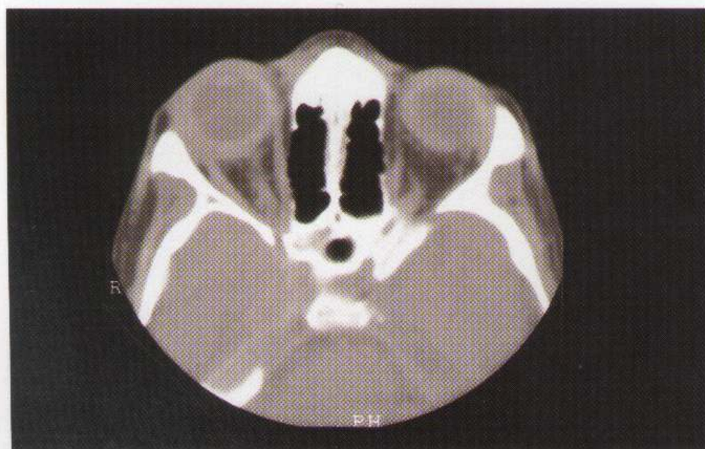


Fig. 7.24
CT scan in right posterior scleritis showing scleral thickening and mild proptosis

detachment, ring choroidal detachments (Fig. 7.21) and subretinal lipid exudation (Fig. 7.22).

3. **Investigations**
   a. *Ultrasonography* shows thickening of the posterior sclera and fluid in Tenon space giving rise to the characteristic 'T' sign. The stem of the T is formed by the optic nerve on its side and the cross bar by the gap containing fluid in sub-Tenon space (Fig. 7.23).
   b. *CT* demonstrates posterior scleral thickening (Fig. 7.24).
4. **Differential diagnosis** includes optic neuritis, rhegmatogenous retinal detachment, choroidal tumour, orbital inflammatory disease or mass, uveal effusion syndrome, Harada disease and intraocular lymphoma.
5. **Treatment** in elderly patients with associated systemic disease is as for necrotizing anterior scleritis. Young patients without associated systemic disease usually respond well to NSAIDs.

Scleral discoloration

Focal discoloration

This is usually seen anterior to the insertions of the horizontal recti and may be caused by the following:

1. **Senile scleral translucency** which is characterized by oval, dark-greyish areas (Fig. 7.25).
2. **Alkaptonuria** may cause brown-black discoloration (ochronosis) at the insertions of horizontal recti and pigmentation of the pinnae.
3. **Haemochromatosis** causes rusty-brown discoloration.
4. **Systemic minocycline** may cause blue-grey paralimbal discoloration, usually denser in the interpalpebral area, possibly due to the photosensitizing properties of the drug. This may be associated with pigmentation of skin, teeth, nails, mucosa, thyroid and bones.
5. **Metallic foreign body**, if long-standing, may produce rust staining.

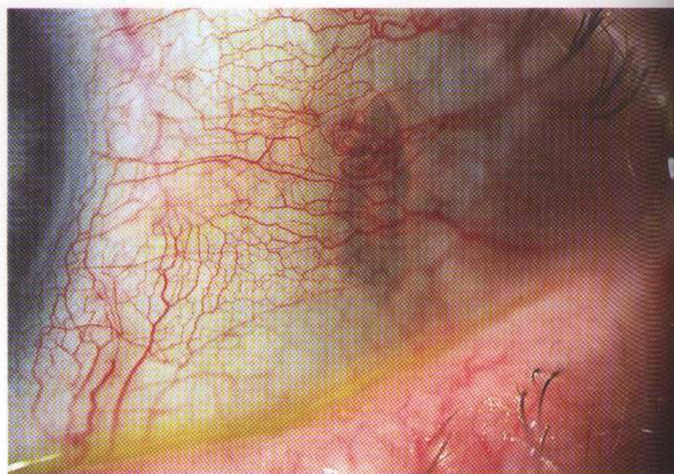


Fig. 7.25
Senile scleral translucency



Fig. 7.26
Blue sclera

Diffuse discoloration

1. **Yellow** discoloration is caused by jaundice.
2. **Blue** discoloration is caused by thinning and transparency of scleral collagen with visualization of the underlying uvea (Fig. 7.26). Important causes include osteogenesis imperfecta types 1 and 2, Ehlers–Danlos syndrome (usually type 6), pseudoxanthoma elasticum (dominant type 2) and Turner syndrome.